ELEPHANTIASIS NON-PARASITICA, SECONDARY TO CHRONIC CARDIAC DISEASE AND REPEATED ŒDEMA OF PREGNANCY.

By T. W. Hastings, M.D.,

INSTRUCTOR IN CLINICAL PATHOLOGY, CORNELL UNIVERSITY MEDICAL COLLEGE, NEW YORK CITY.

An early case of enlargement of the lower limbs to a "most monstrous magnitude" from anasarca, and two cases of obliteration of the thoracic duct, probably without enlargement of the limbs, one from Valsalva (1666–1723) and one from Santorini (1681–1736), are referred to by Morgagni.[1] In 1817 Chevalier[2] wrote of a case of elephantiasis of the right leg following an attack of phlegmasia alba dolens, and in the same year Kennedy[3] contributed an article concerning a patient observed in Madras in 1796.

Kennedy's[3] patient was a male Madrasi, thirty-five years old, who had received six wounds of the left arm from a bursting matchlock. The smaller wounds healed in three months and the larger in four months, during which time much pus was discharged and the head of the ulna came away. The history after the first day was that of an infected wound. The heat, pain, and throbbing were relieved by cold applications, but the enormous swelling, which had gradually extended up his arm, never subsided, and when the wounds had healed the left arm was twice the size of the right. The arm was finally amputated.

Wormi[5] reported a case of enlargement of the left arm consisting of distention of the veins, chronic œdema, thickening of the skin and aponeurosis, and no pus, resulting from complete obstruction and dilatation to double its size of the left subclavian vein from pressure by a yellow, hard, slightly adherent mass at the junction of the subclavian with the jugular. A similar mass pressed upon the descending portion and obliterated the outlet of the thoracic duct, and a bony pelvic tumor by pressure caused extensive œdema of the right thigh. Turner[6] in 1859 reported two cases of complete obliteration of the thoracic duct by pressure from an aneurysm of the aorta, in one of which the area of obliteration extended from the second to the fifth dorsal vertebra, in the other it was opposite the level of the eighth dorsal vertebra.

Not until 1863 was the filaria parasite discovered by Demarquay,[7] and not until 1874 were cases of elephantiasis arabum described by Lewis.[8] In 1867 T. Carr Jackson[9] reported an interesting case of elephantiasis following infection, in which the elephantoid swelling of the left thigh followed the occurrence of a large intraperitoneal "abscess" extending transversely across the lower abdomen. Three days after recovery from the abscess the thigh began to swell and during the succeeding six years the dilated lymph varices over the thigh occasionally ruptured and discharged.

The two excellent monographs by Samuel C. Busey,[10] [11] of New Orleans, published in 1878, included the reports referred to and reports of cases of diseased conditions of the lymph channels observed by himself and published in the *New Orleans Medical and Surgical Journal* during the years from 1876 to 1878 inclusive. Busey's two monographs are classics. In summing up certain cases of non-parasitic elephantiasis Busey[10] concluded that their development depended upon some "congenital defect, from nutritive disturbances and changes taking place during intrauterine life, and some acquired factor such as local inflammation." He mentioned two forms, viz., congenital and acquired. The congenital form, resulting from congenital hypertrophy, occurs in acephalic and other monsters, is local, and affects isolated portions of the body, one or both legs, most frequently the right leg. In later life the immediate cause of enlargement may be an inflammation of the derma involving the bloodvessels and lymphatics or primarily the lymphatics, with effusion; this may result in hypertrophy beginning primarily in the subcutaneous tissue and possibly extending to and involving the other soft tissues (muscles, nerves) and the bony structures. The congenital form is usually of the type elephantiasis mollis of Virchow. The conclusion is inevitable (p. 165) that "blood impoverishment, a meagre diet, and bad hygiene are at least exciting agencies which call into active operation a preexisting predisposition." . . . "These circumstances relate more especially to certain acquired forms; but a large number of these manifestly originating in some congenital abnormality are developed in after life under influences which may be regarded as exciting causes." The age of puberty is regarded by several writers as the period of most prolific development.

In the acquired forms, such as elephantiasis arabum (filariasis) and the condition due to repeated local inflammation and pressure upon large vessels, the tissue changes correspond to true elephantiasis dura of Virchow. Cholmeley[12] in 1869 reported a case dependent upon "obstructed heart circulation and engorgement of lymph channels in parts remote from the direct influence of such impediments," a condition apparently similar to that of the case here reported. A female child, the fourteenth of seventeen children, the sixteen in good health, was born at term, deeply cyanosed. Repeated attacks of dyspnœa and "inflammation of the chest" occurred during a period of from four to five years. The child was always well nourished and fat, with good, bright color in the cheeks, but easily affected by cold, and at such times complained of want of breath and a feeling of tightness in the chest, and the complexion assumed a markedly livid tint. The pulse was "normal in frequency, rhythm, volume, and force," but all over the heart was heard a soft blowing systolic murmur loudest at the junction of the second left costal cartilage with the sternum. "During her sixth year swelling appeared over the right ankle, which gradually extended upward, though not above the knee until two years had elapsed; but in the third year when the patient was between seven and eight, the 'swelling extended slowly and steadily upward till her whole limb was implicated,' but has not gone above 'the inguinal line.' The increase in the size of the swelling was always greater toward evening, and did not affect the foot when a boot was put on when the child first got up."

"The entire limb was uniformly enlarged, felt 'soft, firm, and elastic,' the lower part being firmer and more tense than the upper; in color and temperature the limbs did not differ." . . . "The skin was smooth and soft as far down as the middle third of the leg; below it was 'harsh, rough, dry, and scaly; on the outer surface of the ankle were a number of 'soft, smooth, red, flattened papules,' not larger than a split pea. On the hypertrophied skin of the great, second, and third toes were 'rough, hard elevations looking much like a half-aborted and dried herpetic eruption, from which occasionally a discharge took place,' and over the tendo Achillis was a 'humid patch from which a milky looking alkaline fluid dripped,' similar in character to the fluid which issues through punctures made into the lower part of the limb, which exhibited under the microscope broken-up cells, granular matter, some oil globules, blood corpuscles, and 'coagulated on boiling.' "

The comparative measurements of the lower extremities were as follows:

	Left.	Right.
At the ankle	8 inches	9¾ inches
At the midleg	9 inches	14 inches
Below the knees	9¾ inches	14½ inches
Above the knees	10½ inches	16½ inches
Upper part of thighs	15 inches	17½ inches
(Child 7–8 years old)		

" There was no fulness or swelling of any kind detected in the groin or pelvis; nothing abnormal in the condition of the right nympha or labium; never any pain in the limb; nor any injury, accident, or known cause to account for the condition." Of Cholmeley's case Busey states (pp. 136–138): No postmortem was permitted so we cannot "positively establish any connection between the lymph stasis, which was first apparent near the ankle, and the disturbed cardiac circulation—the cause of which was probably located in the pulmonary artery; and though it seems improbable that an obstructive force originating in the retardation of the blood current from the right ventricle could be transmitted backward along the fluid contents of the thoracic duct and lymph vessels and glands through which the lymph must pass from the lower extremity to reach the receptaculum, yet in view of cases previously cited such explanation of the phenomena cannot be excluded."

Localization of stasis to the one extremity "must, perhaps, find its explanation in some abnormal condition of the lymph vessels of the right lower extremity, which like the cardiac defect was probably congenital . . . The early history of the swelling and engorgement shows that the accumulation was hypostatic. Movement of the fluid, according to relative elevation of the upper and lower parts of the affected limb, demonstrated the insufficiency of the valves."

In 1869 Bryk[13] reported a somewhat similar case, dependent in part upon cardiac disease apparently acquired. A female servant, twenty years old, from her ninth year had had frequently recurring attacks of erysipelas of the left leg and foot, which in her sixteenth year eventuated in the establishment of an ulcer on the inner side of the ankle, followed by swelling and enlargement of the foot and leg, with horny and thorn-like excrescences upon the thickened and callous skin. "The heart was enlarged transversely, the area of dulness extending from the edge of the sternum to beyond the left nipple, and there was a systolic murmur at the apex. The liver extended the width of three fingers below the arch of the ribs; the spleen, as well as the submaxillary and, especially, the left inguinal glands, were enlarged and indurated." Busey[11] (pp. 139 –140) quotes this case and relates that "the history of this case, as well as its course, apparently located the origin of the elephantiatic development in the lymphatic system. In view, however, of facts primarily presented the insufficiency of the mitral valves cannot be wholly excluded as a factor in producing the stasis of the lymph."

These two cases are quoted in detail because their histories and the elephantiatic conditions resemble the case herewith reported, particularly Cholmeley's case, in which there was no inflammatory condition as an exciting cause. Bryk's patient developed elephantiasis after repeated attacks of erysipelas, and the description of the scars on the affected limb suggests a difference in appearance, may be, sufficient to separate the enlargements due in part to infection and enlargements developing without infection. A second case quoted from Bryk[14] tends to substantiate this difference. A blacksmith, thirty years of age, gave a history of good health up to five years ago, when he suffered from erysipelatous attacks which developed in varices about the left knee-joint and later in the entire leg and in the arm. One year later the right leg was the location of an erysipelatous attack with abscess formation upon the leg and in the inguinal glands. After cicatrization of the inflamed parts at first the foot and afterward the leg "began to be thickened elephantiatically." . . . "With the exception of the funnel-shaped scars in the right inguinal region the skin of the thigh was smooth, but in the leg were numerous scars, and in places the surface was uneven and rough on account of the densely crowded, hard nodes the size of nuts, not movable over adjacent parts . . . and furrowed by numerous rugæ passing in every direction." Sensibility was undisturbed. The glands were generally enlarged, but not painful. A strong systolic murmur was perceptible over the femoral artery of both extremities; the thoracic organs were healthy; no albumin was in the urine.

Continuance of bandaging alone reduced the volume, which increased when the bandage was not employed. After ten months the femoral artery was ligated below Poupart's ligament, "with the effect of reducing the limb to its normal dimensions in the course of two months, but another relapse soon followed."

Quincke[15] reported the following case in 1875: A woman of thirty years with "peritoneal dropsy"—effusion of chyle into the peritoneal cavity—developed œdema of the right leg and forearm and elephantiatic formation, probably due to disturbances of lymphatic circulation of the parts affected. The integument was tense, thickened, and the surface irregular; sometimes a clear yellowish fluid dribbled from integumentary fissures. A postmortem revealed "fatty degeneration of the heart," an inflammatory thickening of the mesentery, and cadaveric emphysema.

Of this case Busey says (p. 80): "An œdematous condition and elephantiatic formation of the right leg and forearm due to the mesenteric condition, viz., an inflammatory thickening of both folds of the mesentery and transformation of the interposed adipose tissue into tense connective tissue." This case is quoted to emphasize the possible development of an elephantiasis from circulatory disturbances of the blood and lymph channels, independently of local inflammation in the extremities. A year after Quincke's report Winckel[16] described a case of enlargement of the left lower extremity, similar to phlegmasia alba dolens, tense and painful, without fever, which developed during the existence of chylous ascites, from which was withdrawn by tapping a milky fluid containing filaria.

From 1878 to 1882 there are no good records of such cases in the literature. In 1882 P. zur Nieden[17] published an inaugural dissertation upon a case of lymphangiectasis with lymphorrhagia affecting the labia. His quotations and views in regard to the pathological condition are referred to under the Description of the Pathological Condition.

From 1882 to 1898 the literature contains no good records of non-parasitic elephantiasis. In 1898 Jopson,[18] of Philadelphia, reported two congenital cases, one at four years and one at one and a half years of age, which he considered the result of intrauterine infection and lymphangitis of the limbs. He recognized four types of the congenital condition, viz.:

Elephantiasis congenita lymphangiectatica;
Elephantiasis congenita telangiectatica;
Elephantiasis congenita fibromatosa;
Elephantiasis congenita neuromatodes.

Among the more recent cases are the following: Favarger[19] (1901) reported a case of non-parasitic elephantiasis due in part to a weak heart. The man, a native of Styria, twenty-five years old, was a farmer who had never left his native country. At fifteen years of age he suffered from a left pleural inflammation which recurred during the following two years. Œdema of both feet was noted at eighteen years of age, and later chronic eczema developed. The heart action was weak, the sounds were clear. The swelling and œdema in the left leg increased until they were enormous, and the scrotum, joints, and right leg below the knee were also swollen. Favarger thought this a sporadic case of elephantiasis, without a possibility of filarial invasion, resulting from a predisposition to chronic œdema readily satisfied by the "weak" heart action which maintained a cardiac stasis as well as lymph stasis, and infection with the cocci of erysipelas or other bacteria and subsequent increase in connective tissue.

In 1902 Rolleston[20] reported two cases of moderate enlargement of the legs in two patients, brother and sister, the condition being a peculiar œdema of the tissues which was permanent as long as they led an ordinary life; after exercise it increased, but disappeared after rest in bed for some days. He suggested that the persistent tendency to œdema from gravitation depends upon some inherent defect or peculiarity of the small bloodvessels, which allows of excessive transudation on slight provocation.

Pokrovski[21] (1902) reported two cases of elephantiasis due to syphilis and cited cases of syphilitic elephantiasis described by Minokoff in 1893 and by Orloff in 1901. Tchlenoff[22] in the same year described a case of elephantiasis of the vulva in a syphilitic woman thirty-eight years old, who had had seven miscarriages and had been under treatment for years. He thought some other factor besides syphilis necessary to produce the condition, but made no definite statement in regard to such factors.

A case of elephantiasis dependent upon a congenital defect in the lymph-vessel system of a male child, two years old, was reported by Bernhard and Blumenthal[23] in 1902. The child was admitted to the hospital for a deformity of the external genitalia. Shortly after birth an operation for phimosis was performed, and at this time the elephantiatic condition of the left foot was noted. After operation the prepuce swelled continuously and the entire left leg developed a condition of pronounced elephantiasis, both the external genitalia and the leg presenting a furrowed, greatly thickened skin. The left leg measured from 4 to 8 cm. more than the right in corresponding parts. Surface temperature and sensibility were the same in the left and the right legs, but the surface of the left was drier and harsher on palpation. Microscopic examination of the tissue showed a diffuse fibromatous change in the connective tissue and marked lymphangiectasis. The family history was excellent and there was no history or evidence of an earlier inflammatory condition of the left leg or genitals. They thought the enlargement dependent upon some congenital anomaly of the lymph-vessel system. This case resembles Friedberg's case of colossal development of the right leg, in which, the condition was extremely marked at birth, however, and deformities of the skeletal as well as of soft parts existed.

Cases of elephantiasis have been reported not infrequently in the United States, some of them parasitic in nature and imported from tropical countries where filariasis is endemic. In recent years I have seen four cases of filariasis, early cases with no change in the subcutaneous tissues.

The case here recorded seemed to me to be one having as etiological factors the chronic cardiac condition and some local lymphatic deficiency in the right leg. The history does not make clear the duration of the cardiac disease, but does exclude the probability of parasitic invasion, and the normal blood condition bears out the history. I am aware of the fact that in parasitic elephantiasis filaria embryos are rarely found in the blood. (Manson's *Tropical Diseases,* 1899, p. 466.) Entire freedom from attacks of inflammation in or repeated injuries to the right leg suggested that the local condition depends upon some defect in the lymphatics of the part affected, whether congenital or acquired it is difficult to prove. The occurrence and persistence without steady increase in size of an œdematous condition of the left leg suggest more strongly some local lymph-vessel deficiency in the right leg. The repeated occurrence of œdema during three pregnancies and the cardiac disease with several periods of broken compensation would account for the œdema in both legs, and yet not for the thick, elephantiatic condition of the skin of the right leg. The case corresponds closely to the cases of Cholmeley and Quincke, quoted from Busey's monograph, cases which Busey considered due to latent congenital defect combined with accessory causes later in life, thereby producing an acquired condition depending partly upon a congenital defect.

C. W., a native of the United States, twenty-eight years of age, by occupation a silkwinder, was admitted to the Medical Dispensary, March 13, 1902, complaining of pain in the left arm and left chest.

Family History. Parents are living; mother has suffered from "Bright's Disease" and dropsy" for years; one sister has heart disease and has been under observation at the Dispensary for mitral stenosis; one brother died of consumption at twenty-four years of age; other brothers and sisters died in infancy.

Personal History. The patient was born in Freehold, N. J. When an infant the family moved to Chicago, thence to Philadelphia, thence to New York, eighteen years ago, and the patient has never been out of these four States. No illness occurred in childhood excepting measles. During the last three years she has been questioned occasionally in regard to early illness and important details, and the history given in 1902 is correct. Excepting measles, there is absolutely no history of infectious disease or of local infection. Menstruation was established during the eleventh year, and when seventeen years old she married. Five years after marriage she had been the mother of four children, one of whom died of meningitis, a second of diphtheria, a third at two weeks of age from an undetermined cause, and the fourth is living and in good health. Her husband died of pulmonary tuberculosis six years after their marriage. With each of the four pregnancies there was moderate bilateral œdema of the legs, no more marked in right than in left, but never a permanent swelling of either leg after pregnancy. During no one of the pregnancies were there signs or symptoms of femoral vein thrombosis or inflammation of the legs, and the condition of the limbs above the knee confirmed the statement. There was no fever or illness following parturition. After her husband's death she began work as a silkwinder, which necessitates constant walking to and fro, and within two years' time the right ankle began to swell during the day, the swelling subsiding at night, and gradually over a period of one and a half years the leg below the knee became much enlarged. For four years the permanent swelling has been slowly increasing.

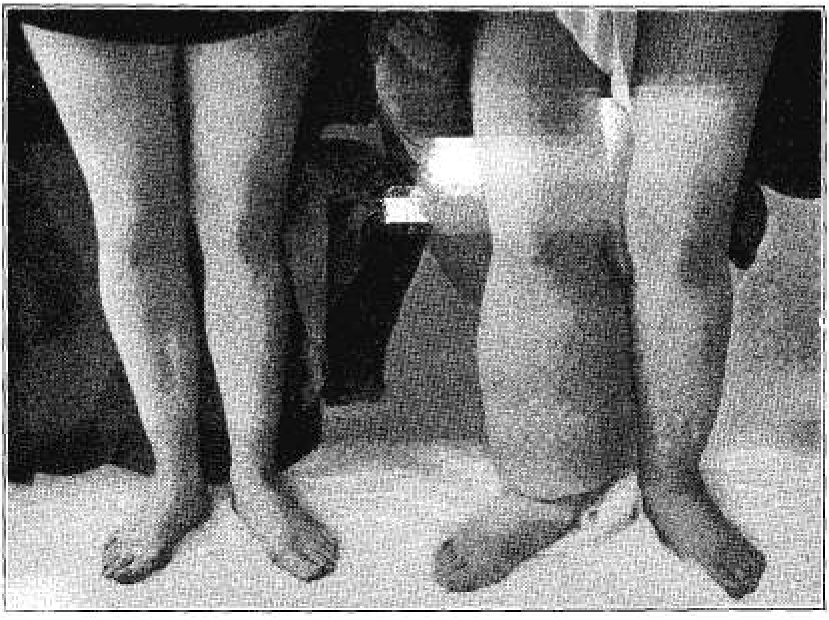
There has been no urinary disturbance and the urine has never been milky.

Present Illness. Four days before admission (March 13, 1902) she was suddenly taken with a sharp, darting pain in the lower left chest, which after from two to three hours extended to the left arm and was sufficiently severe to prevent sleep. There was dyspnœa after exertion. For two weeks the left chest had felt uncomfortable and the left foot had swollen. There was "increased" urination; no cough.

Examination. The lower eyelids were puffy, the pupils dilated, and the eyes deeply dark-ringed. The tongue was lightly coated. Thorax well formed, sternum straight, and lungs normal throughout.

Heart. The apex-beat was visible in the fifth space 10 cm. from the midsternum; the maximum impulse was 11 cm. from the midsternum in the fifth space, well outside the midclavicular line. There was no thrill and the shock of the first sound was felt. The area of relative cardiac dulness was enlarged to the left and extended not quite to the sternal border to the right. Cardiac pulsations were 114 to the minute. At the apex a blowing systolic murmur, soft and of short duration, was heard; in the fifth space, from 11 to 12 cm. from the median line, there was a loud rumbling diastolic murmur, not constant; and mesial to the maximum impulse a loud, high-pitched blowing diastolic murmur was constantly heard; this murmur increased in intensity toward the aortic area, was audible though less distinct at the left of the sternum in the second interspace, and loudest in the third and fourth interspaces to the left of the sternum. The tricuspid area was clear. The second aortic sound was obliterated, the second pulmonic sound was not accentuated. The pulse was full, large, quick, not typically Corrigan. There was a well-marked capillary pulse in the finger nails, and a well-marked Riesman-Müller sign upon examination of the throat.

FIG. 1



To the right, C. W., aged twenty-eight years, the subject of non-parasitic elephantiasis, aortic insufficiency, and rheumatic purpura. To the left, M. A., aged twenty-four years, a sister of C. W., and a subject of mitral stenosis (the legs and thighs normal). Photograph taken March 27, 1902.

The right leg from the knee down was greatly enlarged, measuring 58 cm. in the largest circumference in the middle third of the calf; the enlargement was fairly regular and uniform (Fig. 1). The surface was smooth, with the exception of a pitted area 2 cm. in diameter, suggesting a scar near the lower third anteriorly, and exaggeration of the skin folds near the ankle. Over the posterior surface (Fig. 2) were scattered six small pitted areas, the skin covering which was in no way different from that over the remainder of the leg. The skin was markedly thickened, boggy, but not tense, and did not pit on pressure as did the simple œdematous left leg, nor was the skin rough and harsh as in filarial elephantiasis. There was no local temperature, no swelling, no tenderness. The hair overlying the anterior surface particularly was long and had been "frequently cut off." The patient stated that after standing or walking the right leg was as hard as a rock and the bandage worn about the swollen calf became "saturated with water." Scattered over the surface were a few dilated venules. The left leg presented a moderate œdema and some thickening of the soft parts, and also a pitted scar over the lower third anteriorly. The greatest circumference of the left calf was 34 cm. The œdema existed over both feet and in the left limb up to the lower third of the thigh. The bony structures about the right knee-joint were evidently enlarged and thickened, particularly the condyles of the tibia.

The thickening of the skin did not suggest scleroderma.

Abdominal Examination. Pelvic and abdominal examinations were negative. The cardiac condition was treated, with no change in the size of the right leg, but with partial disappearance of the œdema in the left leg.

The history and the preceding examination were recorded on March 13, 1902. Two weeks later, on March 27, she returned, with a history of pain and soreness across the abdomen for three or four days, some pain in both knees, ankles, and bones of the forearms. There was no chill, no fever, no vomiting or nausea, but sweating and severe headache. There was no bleeding from the nose, mouth or bowel, and the urine had been normal in color. Thirty-four hours before the visit and two days after the onset of pains in the joints she suffered from a sudden attack of intense pain in the left side of the abdomen, and bright-red spots appeared on the thighs and upper legs, most abundant about the knees, also on the dorsal surfaces of the forearms and wrists. On examination the spots were dark purple-red, in size from 1 mm. to a blotch from 2 to 3 cm. in diameter. The knee-joints and inner sides of both thighs were tender. The swelling of the legs had not changed.

The abdomen was tender in the upper zone, otherwise negative. The lungs were normal.

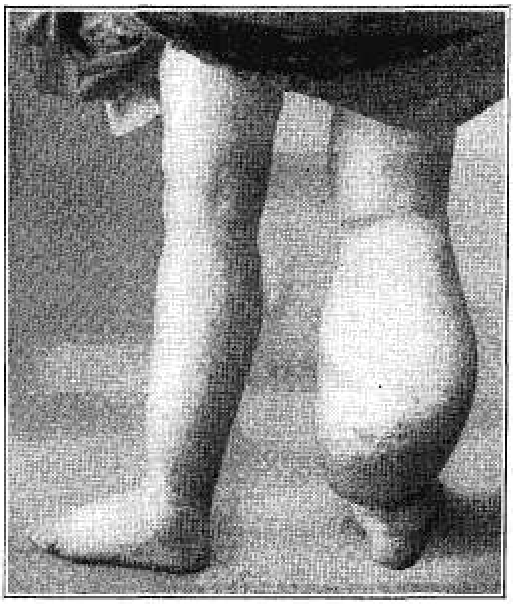
The cardiac area had decreased, the maximum impulse being 9.5 cm. from the midsternum in place of 11 cm. on March 13th; the loud diastolic aortic murmur replaced the second aortic sound and the pulmonic second sound was intensely sharp and accentuated. The pulse was 90 to the minute, quite regular in rhythm and force.

At this visit the photographs were taken, and the purpuric spots are clearly defined (Figs. 1 and 2).

The two blood examinations had shown a normal blood condition, with the exception of a slight increase in the erythrocytes; hemoglobin, 88 per cent.; red blood corpuscles, 5,600,000; white blood corpuscles, 8000. Differential count of the leukocytes (300): polymorphonuclear, 71 per cent.; lymphocytes, 21 per cent.; large mononuclear, 8 per cent.; eosinophiles, 0 per cent.; mast cells, 0 per cent.; parasites not found.

January 19, 1904. The patient returned after an absence of twenty months, with the following history: On June 10, 1903, after a fall, when she bruised the inner side of the enlarged right calf, "gangrene set in." She was taken to a hospital and remained there three months, until September, 1903, since when she has visited the hospital twice weekly to have the last wound dressed.

FIG. 2



Rear view of C. W., non-parasitic elephantiasis.

Three operations were performed under cocaine. At the first "pus" was obtained, but later the discharge became serous, as it is at present. At the time of the operations her medical attendants stated that the skin was one and one-half inches thick.

She complained again, (January 19, 1904) of dyspnœa, aching in the arms and thorax, and of excessive "pounding" of the heart. General health had been excellent. The œdema of the left and enlargement of the right leg had not increased. The cardiac area had again extended, there was a diastolic rumbling murmur at the apex in the fifth interspace from 10 to 11 cm. from the midsternum, and a loud diastolic aortic murmur most intense in the third and fourth spaces along the left sternal border. The tricuspid area was clear. The pulse was large, quick, regular in rhythm. The lungs were normal. Abdominal examination was negative.

The œdema of the left leg about the ankle was moderate. The dressing about the right leg below the knee was wet with a sticky, serous fluid. There was no thickening or dilatation along the veins of the right leg, and nothing abnormal about the genitalia.

The measurements of the two legs were:

	Right.	Left.
Ankle	26 cm.	18 cm.
Two inches below knee	44 cm.	
Three inches above knee	37 cm.	
Largest measurement of calf		36 cm

Above the right knee, in lower third of thigh, the skin was slightly thickened. The left leg was œdematous from knee to ankle, and œdema extended to both feet. The right leg had become much reduced in size, the largest measurement of 44 cm. being 14 cm. less than the 58 cm. of March 13, 1902.

The patient's last visit was in March, 1905, for the cardiac condition, and at that time the operation wound of June, 1903, had not healed, the condition being that of a fistulous opening discharging serous fluid, similar to the conditions of lymphorrhagia or lymphorrhœa recorded by Busey,[10 11] by Fischer,[24] and by P. zur Nieden.[17]

This constant discharge of serous fluid undoubtedly accounts for the reduction in size of the right leg, from 58 cm. in 1902 to 44 cm. in 1904 and 1905. At no time has a milky discharge been noted, although she was requested to look for it. The serous discharge has in no way affected her general health, which is good. She still follows the occupation of silkwinder, and responds readily to treatment when the cardiac condition loses its compensation.

It has not been possible to obtain a section of tissue for histological study. There seems no doubt, however, that the condition is dependent upon the repeated œdema during four pregnancies, leading to dilatation and may be insufficiency of the valves of the lymph vessels, the cardiac disease, with its consequent attacks of stasis, the patient's standing occupation, and possibly some congenital deficiency of the lymph-vessel system of the right lower extremity. History and evidence of infection are wanting, and fortunately infection has not occurred since the operations in 1903.

Aside from bandages properly applied, rest with elevation of the limb when possible, and massage, the treatment has been directed to the cardiac disease.

DESCRIPTION OF THE PATHOLOGICAL CONDITION.

Allard,[4] in a brochure entitled "De l'inflammation des vaisseaux absorbant lymphatique" (Paris, 1824), defined elephantiasis as a "chronic inflammation of the lymphatics of the affected part and ultimately of the remainder of the body."

Busey[10] accepted Virchow's description of the changes in the tissues and recognized an elephantiasis dura, in which a connective tissue of stiff, glistening, white fibers, very firm and almost scirrhous, was found in place of the soft parts; and an elephantiasis mollis, in which the enlarged parts consisted of a soft, gelatinous, uniform tissue. Busey quoted Kaposi as stating that the two changes were not indicative of distinct diseases, and again quoted Kaposi and Tillbury Fox in regard to the fluid found in these tissues, the former considering it a "fibrinogenous" substance, the latter stating it to be lymph.

P. zur Nieden[17] in a monograph considers in detail the genesis of elephantiasis depending upon lymphangiectasis resulting from inflammation. With the initial and prodromal erysipelatous skin inflammation the lymph vessels and glands take part, and thus thrombosis of the lymph vessels and blocking of the glands due to cell-growth accounts for a mechanical lymph stasis. Retention of "tissue juice" is a source of irritation and furnishes material for hyperplasia. Quite the contrary is Wernher's[25] view that the lymph vessels in no place, either capillary or in the glands, are blocked, but the stasis of lymph has ground only in the varicose dilatation, and hereto is the elephantiasis secondary. As a cause for varicosities (lymphatic) Wernher considers gravity the first, as with venous varices. Lallement[26] accepts an atony of the lymph-vessel wall in consequence of repeated erysipelatous attacks as the most important etiological factor. Schliz,[27] after examining the skin from a case of elephantiasis, concluded that the obliteration of lymph vessels of deeper layers occurred through endothelial growth of the walls, and he quotes Virchow[28] as noting the similar endothelial growth in elephantiasis without an explicit statement as to obliteration. Rindfleisch,[29] in a case of pachydermia lymphangiectatica scroti, found a hyperplasia and neoplasia of musculature, and thought the lymph-vessel branches to be compressed by contraction of this muscle. Nieden could not explain the absence of elephantiasis in cases with well-marked lymphangiectasis. Nieden's article contains sixteen careful analyses of lymph from cases of lymphorrhagia.

Jarisch[30] in *Nothnagel's System* describes elephantiasis as resulting from a chronic circulatory disturbance particularly of the lymph system or thoracic duct, accompanying and beginning in acute inflammation and resulting in hyperplasia of the subcutaneous connective tissue. The condition (p. 839) often depends upon recurrent erysipeloid inflammations, phlebitis from lupus, syphilitic ulcerations, ulcers of the foot, and filarial invasion. All of these acquired forms are classified under elephantiasis arabum and separated from the congenital forms, viz., elephantiasis telangiectatica, elephantiasis lymphangiectatica, and the neurofibromatous conditions. The acquired elephantoid condition occurring without inflammation is not mentioned.

Neisser and Jadassohn[31] state that with venous stasis there occur purulent inflammation and connective-tissue hyperplasia of the lymph-vessel system, resulting in the condition called elephantiasis, particularly after recurrent erysipelas (p. 74). Often a true elephantiasis follows a ring-ulcer of the leg. Three forms are recognized, all consisting of a chronic hyperplasia of subcutaneous connective tissue, with more or less œdema and venous hyperæmia:

(*a*) Elephantiasis nostras;

(*b*) Elephantiasis of tropical regions;

(*c*) Elephantiasis-like forms of connective-tissue tumor occurring as congenital deformities in contrast to the acquired forms (*a*) and (*b*).

Elephantiasis nostras results from a chronic or recurrent inflammation combined with stasis in the lymph- and bloodvessels occurring with chronic eczema, chronic ulcerations, lupus, syphilitic tumors, phlebitis, lymphangitis, and particularly with erysipelas due to the streptococcus (Bockardt). Sometimes (p. 227) chronic œdema without history (carefully taken) of earlier erysipelas (although erysipelas may be afebrile and mild; Reclus' "Halzphlegmonie" equals bacterial infection without fever) may be the apparent cause; and more rarely the condition may be postoperative or neuropathic, with neuritis or paraplegia. This type (*a*) affects usually the legs; more rarely the arms, scrotum, labia majora, and the face in tuberculous and syphilitic conditions. There is also a form (p. 229), not to be confused with (*c*), which is congenital in origin, due to intrauterine streptococcus infection of the fœtus from an erysipelatous mother.

The tropical form (*b*) is due to Filaria humani. The group (*c*) of congenital connective-tissue tumors, or tumors having a congenital "grundlage," best called fibromas, are often boggy and tumor-like thickenings of the skin, of which three types are to be recognized:

Elephantiasis telangiectodes;

Elephantiasis lymphangiectodes;

Elephantiasis with involvement of the peripheral nerves, equal neurofibroma. ("Dystrophie œdémateuse héréditaire" and "trophœdéme chronique héréditaire".)

Clinically (p. 227) the four following conditions are to be recognized:

Elephantiasis glabra (smooth);

Elephantiasis nigra (spotted and dark);

Elephantiasis verrucosa sive papillaris;

Elephantiasis lymphangiectodes (with lymphorrhœa).

Fischer's[24] monograph in one part (1901) discusses thoroughly all phases of disease of the lymph vessels, and for a complete bibliography the reader is referred to his publication (pp. x–xiv). At the end of the eighteenth century the normal anatomy of the lymph-vessel system had been carefully described by Henson, Mascagni, and Cruikshank; and Assalini and Sömmering had explained disease processes on the theory of interruption to the flow of lymph through the lymph-vessel system. Later, Allard and Andral concluded the disease processes to be due to inflammation, and this view was upheld by the French school—Velpeau, Breschet, J. Roux, Chassaignac.

During the last eighty years acute inflammation, tuberculosis, and malignant tumors of the lymph vessels have been reported. Fischer separates the acute infectious conditions into two groups, *A* and *B:*

A. Lymphangitis superficialis,

1. Lymphangitis truncularis (ascendens).

(*a*) Simple acute form;

(*b*) Purulent subacute, involving the larger collected bundles of vessels, in surgical blood poisoning and glanders.

2. Lymphangitis reticularis, involving the net-work of vessels in the skin: subepidermal, dermal, or subdermal lymph vessels, conditions often not distinguishable from erysipelas, although they develop no indurated line of demarcation as does erysipelas; and

3. Lymphangitis gangrenosa.

B. Lymphangitis profunda, involving the deeper tissues.

The chronic inflammatory conditions are not specific but result from failure to destroy the source of irritation from an old infection in ulcers and chronic inflammations leading to recurrent lymphangitis. Fischer recognizes two types, viz.:

(*a*) Reticularis (chronic);

(*b*) Truncularis (chronic).

The course of chronic inflammation of large vessel trunks is without pain. The lymph circulation of the area affected is hindered and the collateral circulation is not in function; this condition results in an œdematous swelling peripheral to the area in which the lymph vessels are blocked, sometimes with lymphangiectasis. Particularly if deprived of treatment are the cases of "Strang" (ascending), lymphangitis prone to result in "elephantiatic" changes, since the walls of the vessels, closed by endothelial hyperplasia or by thrombi, undergo a fibrous connective-tissue degeneration which involves the neighboring and enclosed connective tissue and the smaller vessels, particularly the smaller veins.

Dilatation of the lymph channels, *i. e.*, lymphangiectasis, and the formation of lymph varices and lymphangiomas are often congenital afflictions, appearing with birth or soon after birth. (Fischer quotes Patterson and Busey.) The development of lymphangiectasis may occur later in life and these cases fall into two groups, separated etiologically into

A. The tropical, due to invasion of the lymph channels by filaria sanguinis;

B. The non-parasitic, spontaneous, or after occlusion of lymph channels.

This group of cases has its origin in an external or internal obstruction to the on-flow of lymph from the distal portion of a part, in consequence of which the lymph-vessel content is under abnormally high pressure, so that a bulging of the wall occurs. Interruption of the stream can give rise to contracting pathological changes resulting in marked narrowing of the lumen.

Repeated (recurrent) lymphangitis and erysipelas lead to thrombosis and complete occlusion; or complete occlusion may result from neoplasm of parts (cancer, Richet), or from endothelial hyperplasia (C. O. Weber), or from chronic infection (tuberculosis, Goupil), or from invasion by parasites (Filaria, Lewis, Mazéa-Azema, Scheube). Occlusion may result from external pressure after injury and inflammation which in healing have left a contracting, shrivelling, extending scar (Patterson, Klebs); from tumors gradually increasing in volume in the neighborhood of vessels (Busey); in extensive ascites (Know and Egger). Destruction of lymph-vessel area from acute and chronic suppuration of lymph glands, after extirpation of glands, trauma directly affecting the vessels, as in fracture, extensive section of the soft parts (Trélat, Ledderhose) can lead to obstruction to on-flow and subsequently lymphangiectasis. To other authors the difficulty of reflux of lymph into the venous system, arising from abnormally high pressure in consequence of heart disease, congenital stenosis of the venous ostia, and other obstructions to the circulation, suffices to explain the development of dilatation. In such cases the dilatation is confined to the thoracic duct and the large trunks of the thorax and abdomen (Rokitansky, Löschner), or more rarely the peripheral lymph vessels may also become dilated, particularly those of the thigh and genitals (Petters, Simon). Wright believes that the development of lymph varices may depend upon an abnormal increase of vessel content, without previous inflammation of vessels or glands. Bean and Emmert consider paralysis of the muscularis of the larger lymph vessels a possible cause of dilatation. Unna considers the lymph-vessel dilatation the result of two factors, a hindrance to lymph-flow and to blood-flow, the latter being a pressure which works against the natural blood stream, leading particularly to stasis of neighboring veins. Stasis and retention of lymph do not always lead to dilatation, for collateral circulation may be good. Experimental stasis of ductus and larger vessels does not always lead to dilatation (Magendie, Dupuytren, Conheim); obliteration of the vessels of an extremity does not result in lymphangiectasis (v. Lesser); nor does dilatation often follow extirpation of glands. Accordingly, Langhans, Torock, and Nasse take the view that proliferation processes in the lymph-vessel walls play the greatest part in the production of dilatation.

Fischer does not lay much stress on proliferation processes, but takes the view of v. Esmarch, Kulenkampf, and others, that lymph varices arise only when vessels over a large area are obstructed, are overfilled, and the wall elasticity is thus injured and the valves become inefficient, either from dilatation alone or from inflammatory changes.

One may compare these with venous varices and accept a congenital "anlage," which is to be looked for in an abnormal width and flabbiness of the lymph-vessel walls, which readily lend themselves to the development of dilatation through stasis.

The two forms of lymphangiectasis, (1) reticularis, and (2) truncularis, often occur together.

1. Lymphangitis reticularis consists in a dilatation of vessels and capillaries (and "Wurzelgebiete") leading to a "pachydermia lymphangiectatica," in which the outer epidermis is the primary site, the hypertrophic process in the cutaneous and subcutaneous tissues resulting secondarily from the irritation arising from the primary lymph stasis (Kulenkampf, v. Winiwarter, A. Fischer, Teichmann).

On the normal thickened, sometimes pigmented, skin one finds small blebs, which, after being emptied of their fluid contents, have a sharply defined border. On section one sees close under the papillary layer endothelial cell spaces and widened papillary lymph spaces communicating with a net-work of dilated lymph vessels in the skin and subcutaneous tissue. The "spaces" extend to the point of the papillæ and are separated from a thin epidermal by a thin connective-tissue layer, in some more extensively dilated; the connective tissue is atrophied so that the endothelium is in contact with the epidermal layer; the content of the spaces is usually fluid, sometimes with finely granular net-work masses—the coagulated lymph—which contains the few nucleated lymphoid cells. The sebaceous and sweat glands and hair follicles as well as the papillæ are compressed, whereby atrophy of these structures arises. Bloodvessels completely unchanged or dilated project as bulgings into the spaces; through rupture of these vessels the contents became sanguineous. In the papillary and subcutaneous layers irritation is evidenced by a variable high grade of round-cell infiltration.

2. In lymphangitis truncularis the vessels above and below the fascia of the part are involved; either small sections of a trunk are partially dilated, or the dilatation extends along the whole length in a cylindrical or serpentine form. Changes in the vessel wall at the dilated area show lack of growth of the muscularis (Anger). Nieden states the contrary, but the musculature is strong in growth in the slightly dilated portions; clear statements are wanting in regard to the relations of endothelium, intima, and elastic fibers. The number of valves is lessened, the length of those present reduced and insufficient, whereby the turgescence of the varices on standing and walking occurs without hindrance. The widening, bulging, and thickening of the vessel walls occur most often without symptoms.

The dilated lymph vessels at times can be rolled under the fingers. Often lymphorrhagia or lymphorrhœa is the first symptom or sign, the fluid amounting to a few grams or many pounds in a day. Pressure below a fistula in such a case, on standing, increases the flow. The output depends in part upon the opening, often small and fine (Klebs and Tillmann), or may be a flow or exudation from between the epithelial cells (as in the case here reported) without a fistulous opening, and again large fistulæ which admit a sound and communicate with large spaces. Thus one may find normal skin, or more or less pigmented, hypertrophic, elephantiatic changes in the skin, with small flaxseed or pea-sized simple or confluent blebs which contain clear, milky, or sanguineous fluid. The fluid is lymph and related to chyle (Neiden, v. Esmarch, Kulenkampf). Excessive loss of it may lead to emaciation, weakness, and death. Munk and Rosenstein reported a case with a fistula on the inner side of an elephantiatic left leg, which in hunger discharged a clear, slightly opalescent fluid, and after digestion of a fat meal a milky fat-droplet chyle. These varices of lymph vessel net-work may extend over an entire extremity; in such cases hypertrophic changes in the subcutaneous connective tissue result in elephantiasis (v. Winiwarter).

I am indebted to Dr. E. S. Loiseaux, of Morristown, N. J., for the following account of the condition found at operation.

"S. W. was admitted to the hospital with a subcutaneous abscess in the right leg posteriorly. The abscess was secondary to a skin abrasion, received over its site two weeks previously to admission. On incision the skin itself was very much thickened and the pus cavity was several inches in diameter and two or three inches deep, possibly a little more.

This cavity seemed to be immediately beneath the skin proper, in the superficial fascia—I could not say that it extended down to the deep fascia. The skin itself was about three-eighths of an inch thick and its toughness kept the abscess from spontaneous evacuation.

As far as the hypertrophy is concerned I should say that it was nearly all in the superficial fascia or subcutaneous tissue. . . I considered the whole condition one of lymphœdema, as it appeared in the puerperal period. At this time the blood was examined night and day for filaria and none were found."

With Dr. Loiseaux I believe the condition dependent in part upon "lymphœdema" of pregnancy, and the subsequent hypertrophy of the skin and subcutaneous tissue, in the right leg and not in the left, due to chronic lymph stasis dependent upon lymph-vessel deficiency, probably congenital, as suggested by the earlier observers quoted in this article.

Unfortunately it has not been possible to obtain tissue for microscopic examination.

BIBLIOGRAPHY.

1. Morgagni. De sedibus et causis morborum, Liber ii, p. 152, Venice, 1761; Liber iii, 26. Translation by Alexander, London, 1769, p. 300.
2. Chevalier. Medical and Chirurgical Transactions, London, 1817, vol. ii, p. 63.
3. Kennedy. Edinburgh Medical and Surgical Journal, 1817, vol. xiii, p. 54.
4. Quoted by Hasse: "Diseases of Organs of Circulation and Respiration." Translation by Swaine, Sydenham Society, London, 1846, p. 7.
5. Wormi. Gazette Hebdomadaire, vol. vi, p. 279, 1859.
6. Turner. Edinburgh Medical Journal, 1859, p. 1003.
7. Demarquay. Gazette Medicale, Paris, 1863, xviii, p. 665.
8. T. R. Lewis. "The Pathological Significance of Nematode Hematozoa," Calcutta, 1874.
9. Jackson. Transactions of the Pathological Society of London, 1866, xvii, 287.
10. Busey. "Congenital Occlusion and Dilatation of Lymph Channels, 1878, p. 165.
11. Busey. "Narrowing, Occlusion, and Dilatation of Lymph Channels, Acquired Forms," 1878.
12. Cholmeley. Transactions of Clinical Society of London, 1869, vol. ii, 116. Quoted by Busey, p. 136.
13. Bryk. Oesterreich, Zeit. f. Heilkunde, 1869, 469.
14. Bryk. Ibid., 449.
15. Quincke. Deutsche Archiv f. klin. Med., 1875, xvi, 128.
16. Winckel. Ibid., 1876, xvii, 303.
17. P. zur Nieden. "Ueber einen Fall von Lymphangiectasie mit Lymphorrhagie," Berlin, 1882.
18. Jopson. Archives of Pediatrics, 1898, 174.
19. Favarger. Wiener klinische Wochenschrift, December 26, 1901.
20. Rolleston. Lancet, September 20, 1902.
21. Pokrovski. Medicinskoie Obozrenie, 1902, lvii, No. 8, from Philadelphia Medical Journal, November 8, 1902, 570.
22. Tchlenoff. Medicinskoie Obozrenie, 1902, lvii, No. 11, 859, from Philadelphia Medical Journal, December 27, 1902, 1039.
23. Bernhard and Blumenthal. Deutsche med. Wochen., December 11, 1902, p. 902.
24. Fischer. Deutsche Chirurgie, 1901, Lief. 24 a.
25. Wernher. Deutsches Zeit. f. Chirurgie, 1875, v, 429.
26. Lallement. Cited by Schliz, Archiv f. Heilkunde, 1874, xv, 153.
27. Schliz. Loc cit., p. 163.
28. Virchow. "Die Krankhaften Geschwülste," Band i, 303.
29. Rindfleisch. Lehrbuch der Pathologische Gewebesehre, 1871, 2 auf. 277.
30. Nothnagel's Specielle Pathologie und Therapie, 1900, Band xxiv, 714.
31. Neisser and Jadassohn. Ebstein und Schwalbe, Handbuch der Praktischen Medizin, 1901, iii, 29.
32. v. Winiwarter. Deutsches Chirurgie, Lief. xxiii, 355-402.

RESULTS OF THE BITERMINAL TRANSPLANTATION OF VEINS.

By Alexis Carrel, M.D.,

AND

C. C. Guthrie, M.D.,

OF CHICAGO, ILLINOIS.

(From the Hull Physiological Laboratory, University of Chicago.)

The first series of experiments performed demonstrated that by interposing between the two cut ends of an artery a segment of vein it is possible to restore quickly the arterial circulation, and that the venous segment is able to perform the principal arterial functions. A summary of the immediate results of this operation,